PP-104

High Alkaline Phosphatase Post Kasai Procedure, Should We Look Beyond the Liver?

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INTRODUCTION
Metabolic bone disease (MBD) is a recognized complication in patients post Kasai procedure. In biliary atresia (BA), metabolic disturbance is caused by impairment of the passage of bile salts into the alimentary canal causing inadequate emulsification of fat and thus incomplete absorption of vitamin Progressively, liver cirrhosis develops in BA leading to further impairment of hydroxylation of vitamin D.

METHODOLOGY
We report a 5-year, 7-month-old girl with post Kasai procedure done successfully at Day 71 of life for Biliary Atresia with incidental increase in ALP during routine clinic review. Unfortunately no serial liver function was done as she had defaulted follow up. She did not sustain any fractures before. Clinically she was pink and not jaundiced. She was not clubbed but small for age. Per abdominal examination yield no significant findings apart from healed rooftop scar. Her wrists were swollen. No rachitic rosary noted.

RESULTS
Full blood count revealed Hemoglobin (Hb) 13.4 g/dL and platelets of 329 10^3/UL. Liver function test revealed Alanine Aminotransferase (ALT) 26.31 U/L, Aspartate aminotransferase (AST) 61.77 U/L, alkaline phosphate (ALP) 6996 U/Total serum bilirubin was 5.95 umol/Direct bilirubin of 3.5 umol/L Gamma-Glutamyl-transferase (GGT) was 134 U/Coagulation profile revealed INR 0.96 and APTT 50.8 sec. Vitamin D level unfortunately was rejected and intact Parathyroid hormone was 63.6 pg/mL. Hepatobiliary Ultrasonography showed no biliary tree obstruction with cirrhotic liver. No nodular lesion. We started her on oral calcitriol 1 mcg once daily. The ALP begins to decrease from 6996 U/L to 800 U/L within two months. She has been well during outpatient review.

CONCLUSION
Vitamin D is essential for the bone growth and development. Sustained vitamin D deficiency in children may cause bone deformity, pain, or pathological fractures (i.e., rickets). The management of Vitamin D deficiency include oral vitamin D supplement and pamidronate (bisphosphonates). Clinicians need to remain vigilant in monitoring for MBD especially in patients post liver surgery.

PP-105

The Eyes and Skin as the Windows to the Brain

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INTRODUCTION
The pituitary glands and the eyes stem from the same embryonic origin, which is the anterior neural ridge. Hence, various ocular malformations are reported in the presence of hypopituitarism. Aplasia cutis congenita, on the other hand, is characterized by partial or complete absence of the skin as a result of failure in ectodermal fusion. It can occur as a constellation of ocular involvement, skin lesion and cerebral malformations. We aim to report on a case of aplasia cutis congenita with microphthalmia and coloboma, and subsequently diagnosed as hypopituitarism.

METHODOLOGY
A baby girl was delivered at full term via emergency LSCS due to poor progress of labour. Ventriculomegaly and absence of corpus callosum were detected antenatally. Clinically, there was a defect over left side of her forehead and temporal scalp. The lesion, measuring 7 cm x 3 cm, was longitudinal in shape, erythematous with visible membranous-like structure underneath. Ophthalmology assessment revealed left microphthalmia and optic disc coloboma at the right eye. MRI brain showed atrophied left globe associated with colpocephaly, callosal agenesis and cerebellar hypoplasia. Pituitary glands appeared to be normal.

RESULTS
Her thyroid function test at day 5 of life (T4 9.34 pmol/L/ TSH 2.23 mIU/L) was suggestive of hypothyroidism. L-thyroxine was started. At day 7 of life, she developed hypernatraemia (serum sodium ranged 151-156 mmol/l). Her urine osmolarity (89 mOsm/kg) and serum osmolarity (324 mOsm/kg) were suggestive of diabetes insipidus. The biochemical abnormality resolved with desmopressin. Despite euglycaemia and normal blood pressure, her serum cortisol was low (87.7 nmol/l). She was started on hydrocortisone. She was discharged well and will be reviewed in our outpatient clinic.

CONCLUSION
Current literature has yet to report on the association between aplasia cutis congenita and hypopituitarism. In the presence of other malformations, in particularly ocular involvement, it will be worth screening for pituitary insufficiencies for early detection and intervention.